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Search Results -

Terms	Documents
alpha N-acetylglucosaminidase	4

Database: US Patents Full-Text Database; US Pre-Grant Publication Full-Text Database; JPO Abstracts Database; EPO Abstracts Database; Derwent World Patents Index; IBM Technical Disclosure Bulletins

Search: L1

Refine Search | Recall Text | Clear

Search History

DATE: Sunday, December 01, 2002 Printable Copy Create Case

Set Name side by side	Query	Hit Count	Set Name result set
DB=USPT,PGPB,JPAB,EPAB,DWPI; PLUR=YES; OP=ADJ			
L1	alpha N-acetylglucosaminidase	4	L1

END OF SEARCH HISTORY

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Search Results - Record(s) 1 through 5 of 5 returned.

☐ 1. Document ID: WO 9719177 A1

L2: Entry 1 of 5 File: EPAB May 29, 1997

PUB-NO: WO0009719177A1
DOCUMENT-IDENTIFIER: WO 9719177 A1
TITLE: SYNTHETIC MAMMALIAN alpha -N-ACETYLGLUCOSAMINIDASE AND GENETIC SEQUENCES ENCODING SAME

Full | Title | Citation | Front | Review | Classification | Date | Reference | Sequences | Attachments | Claims | KWIC | Draw Desc | Image

☐ 2. Document ID: WO 2003057138 A2 US 20030143669 A1

L2: Entry 2 of 5 File: DWPI Jul 17, 2003

DERWENT-ACC-NO: 2003-577498
DERWENT-WEEK: 200354

TITLE: Producing a lysosomal hydrolase having an oligosaccharide modified with N-acetylglucosamine-1-phosphate, for treating lysosomal storage disease, comprises expressing a lysosomal hydrolase in a furin-deficient mammalian cell

Full | Title | Citation | Front | Review | Classification | Date | Reference | Sequences | Attachments | Claims | KWIC | Draw Desc | Image

☐ 3. Document ID: WO 200222157 A2 AU 200195028 A

L2: Entry 3 of 5 File: DWPI Mar 21, 2002

DERWENT-ACC-NO: 2002-471182
DERWENT-WEEK: 200251

TITLE: Administering a polypeptide to patient involves introducing aqueous solution of collagen comprising in suspension, a population of cultured vertebrate cells that express the polypeptide, and several microcarriers

Full | Title | Citation | Front | Review | Classification | Date | Reference | Sequences | Attachments | Claims | KWIC | Draw Desc | Image

☐ 4. Document ID: US 20030148460 A1 WO 200119955 A2 AU 200073303 A US 20020025550 A1 EP 1224266 A2 BR 200014514 A US 20020150981 A1 JP 2003509043 W US 6534300 B1 US 6537785 B1

L2: Entry 4 of 5 File: DWPI Aug 7, 2003

DERWENT-ACC-NO: 2001-290356
DERWENT-WEEK: 200358

TITLE: Novel N-acetylglucosamine-1-phosphotransferase and N-acetylglucosamine-1-phosphodiester alpha-N-Acetylglucosaminidase, useful for producing phosphorylated lysosomal hydrolase for treating lysosomal storage diseases

Full | Title | Citation | Front | Review | Classification | Date | Reference | Sequences | Attachments | Claims | KWIC | Draw Desc | Image

☐ 5. Document ID: US 20030039643 A1 WO 9719177 A1 AU 9676124 A EP 870036 A1 JP 2000500972 W AU 720778 B US 6255096 B1

L2: Entry 5 of 5 File: DWPI Feb 27, 2003

DERWENT-ACC-NO: 1997-298114
DERWENT-WEEK: 200318

TITLE: Nucleic acid encoding mammalian alpha-N-acetyl:glucosaminidase - used for the diagnosis and treatment of muco:poly:saccharidosis type IIIB, also used in gene therapy

Full | Title | Citation | Front | Review | Classification | Date | Reference | Sequences | Attachments | Claims | KWIC | Draw Desc | Image

Generate Collection | Print

Terms	Documents
alpha- N-acetylglucosaminidase	5

Display Format: - Change Format

Previous Page Next Page

WEST

Generate Collection | Print

Search Results - Record(s) 1 through 4 of 4 returned.

☐ 1. Document ID: WO 9719177 A1

L1: Entry 1 of 4 File: EPAB May 29, 1997

PUB-NO: WO009719177A1
DOCUMENT-IDENTIFIER: WO 9719177 A1
TITLE: SYNTHETIC MAMMALIAN alpha -N-ACETYLGLUCOSAMINIDASE AND GENETIC SEQUENCES ENCODING SAME

Full | Title | Citation | Front | Review | Classification | Date | Reference | Sequences | Attachments | Claims | KWIC | Draw Desc | Image

☐ 2. Document ID: WO 200222157 A2 AU 200195028 A

L1: Entry 2 of 4 File: DWPI Mar 21, 2002

DERWENT-ACC-NO: 2002-471182
DERWENT-WEEK: 200251
COPYRIGHT 2002 DERWENT INFORMATION LTD

TITLE: Administering a polypeptide to patient involves introducing aqueous solution of collagen comprising in suspension, a population of cultured vertebrate cells that express the polypeptide, and several microcarriers

Full | Title | Citation | Front | Review | Classification | Date | Reference | Sequences | Attachments | Claims | KWIC | Draw Desc | Image

☐ 3. Document ID: US 20020150981 A1 WO 200119955 A2 AU 200073303 A US 20020025550 A1 EP 1224266 A2 BR 200014514 A

L1: Entry 3 of 4 File: DWPI Oct 17, 2002

DERWENT-ACC-NO: 2001-290356
DERWENT-WEEK: 200270
COPYRIGHT 2002 DERWENT INFORMATION LTD

TITLE: Novel N-acetylglucosamine-1-phosphotransferase and N-acetylglucosamine-1-phosphodiester alpha-N-Acetylglucosaminidase, useful for producing phosphorylated lysosomal hydrolase for treating lysosomal storage diseases

Full | Title | Citation | Front | Review | Classification | Date | Reference | Sequences | Attachments | Claims | KWIC | Draw Desc | Image

☐ 4. Document ID: WO 9719177 A1 US 6255096 B1 AU 9676124 A EP 870036 A1 JP 2000500972 W AU 720778 B

L1: Entry 4 of 4 File: DWPI May 29, 1997

DERWENT-ACC-NO: 1997-298114
DERWENT-WEEK: 200140

TITLE: Nucleic acid encoding mammalian alpha-N-acetyl:glucosaminidase - used for the diagnosis and treatment of muco:poly:saccharidosis type IIIB, also used in gene therapy

Generate Collection | Print

Terms	Documents
alpha N-acetylglucosaminidase	4

Display Format: - Change Format

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=> d his

     (FILE 'HOME' ENTERED AT 09:27:57 ON 01 DEC 2002)

     INDEX 'ADISALERTS, ADISINSIGHT, ADISNEWS, AGRICOLA, ANABSTR, AQUASCI,
     BIOBUSINESS, BIOCOMMERCE, BIOSIS, BIOTECHABS, BIOTECHDS, BIOTECHNO, CABA,
     CANCERLIT, CAPLUS, CEABA-VTB, CEN, CIN, CONFSCI, CROPB, CROPU, DDFB,
     DDFU, DGENE, DRUGB, DRUGLAUNCH, DRUGMONOG2, ...' ENTERED AT 09:28:05 ON
     01 DEC 2002
               SEA ALPHA-N-ACETYLGLUCOSAMINIDASE
              ---------
                1   FILE AGRICOLA
                1   FILE ANABSTR
                2   FILE AQUASCI
               87   FILE BIOSIS
                6   FILE BIOTECHABS
                6   FILE BIOTECHDS
               48   FILE BIOTECHNO
                2   FILE CABA
               14   FILE CANCERLIT
              118   FILE CAPLUS
                1   FILE CIN
                6   FILE DDFB
               35   FILE DGENE
                6   FILE DRUGB
              119   FILE EMBASE
               28   FILE ESBIOBASE
                1   FILE FEDRIP
              140   FILE GENBANK
                4   FILE IFIPAT
                4   FILE JICST-EPLUS
               22   FILE LIFESCI
               89   FILE MEDLINE
               45   FILE PASCAL
                1   FILE PHIN
                1   FILE PROMT
               66   FILE SCISEARCH
               29   FILE TOXCENTER
               33   FILE USPATFULL
                6   FILE WPIDS
                6   FILE WPINDEX
L1              QUE ALPHA-N-ACETYLGLUCOSAMINIDASE
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     FILE 'CAPLUS, EMBASE, MEDLINE, BIOSIS, SCISEARCH, BIOTECHNO, PASCAL'
     ENTERED AT 09:29:47 ON 01 DEC 2002
L2           93 S L1 AND (VARIANT OR MUTANT OR DERIVATIVE)
L3           26 S L2 AND PY<1995
L4            4 S L3 AND PURIF?
L5            4 DUP REM L4 (0 DUPLICATES REMOVED)
L6           17 DUP REM L3 (9 DUPLICATES REMOVED)
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antiserum to react with normal, **mutant**, monomeric and multimeric forms of the enzyme.

L5 ANSWER 3 OF 4 CAPLUS COPYRIGHT 2002 ACS
ACCESSION NUMBER: 1976:103375 CAPLUS
DOCUMENT NUMBER: 84:103375
TITLE: Sanfilippo disease type B: presence of material cross reacting with antibodies against .**alpha**.-**N-acetylglucosaminidase**
AUTHOR(S): Von Figura, Kurt; Kresse, Hans
CORPORATE SOURCE: Inst. Physiol. Chem., Univ. Muenster, Muenster, Ger.
SOURCE: Eur. J. Biochem. (**1976**), 61(2), 581-8
CODEN: EJBCAI
DOCUMENT TYPE: Journal
LANGUAGE: English
AB .**alpha**.-**N-acetylglucosaminidase** (I) (enzyme deficient in Sanfilippo disease type B) was **purified** from normal human urine. An antiserum was raised in rabbits against the **purified** enzyme. Preincubation of the antiserum with crude I from normal human urine, followed by centrifugation, led to a marked redn. of the I activity in the supernatant. Formation of the antibody-enzyme complex had no influence on the activity. The thermal stability of the enzyme was markedly enhanced by complex formation with the antiserum. In the urine from three patients with Sanfilippo disease type B the presence of cross-reacting material could be demonstrated by incubating the antiserum with I in the presence of Sanfilippo B urine or by pretreatment of the antiserum with Sanfilippo B urine. Immunodiffusion and immunoelectrophoresis of crude normal or Sanfilippo B urine gave rise to up to four pptn. lines, only one of which exhibited I activity in the case of normal urine. **Purified** I yielded only a single pptn. line. After adsorption with the **purified** enzyme the antiserum did not cross react with any of the urinary proteins. On a quant. detn. of cross-reacting material using Sepharose-immobilized antibodies in the urine from two Sanfilippo B patients the amt. of cross-reacting material appeared to be less than one fourth of the amt. of protein in an age-matched control urine. The cross-reacting material present in the urine of Sanfilippo B patients had a significantly lower binding affinity for antibodies against I than prepns. from normal human urine. It could be calcd. that the amt. of cross-reacting material in the urine of Sanfilippo B patients exceeded that of normal controls. It is concluded that Sanfilippo disease type B is due to a mutation of a structural gene coding for I. The mutation affects the catalytic and immunol. properties of the enzyme protein.

L5 ANSWER 4 OF 4 MEDLINE
ACCESSION NUMBER: 75054959 MEDLINE
DOCUMENT NUMBER: 75054959 PubMed ID: 4215452
TITLE: Physical properties and biological activities of two forms of **alpha-N-acetylglucosaminidase** from bovine spleen.
AUTHOR: Mersmann G; von Figura K; Buddecke E
SOURCE: BIOCHIMICA ET BIOPHYSICA ACTA, **(1974 Sep 11)** 364 (1) 88-96.
Journal code: 0217513. ISSN: 0006-3002.
PUB. COUNTRY: Netherlands
DOCUMENT TYPE: Journal; Article; (JOURNAL ARTICLE)
LANGUAGE: English
FILE SEGMENT: Priority Journals
ENTRY MONTH: 197503
ENTRY DATE: Entered STN: 19900310
Last Updated on STN: 19970203
Entered Medline: 19750319

L6 ANSWER 6 OF 17 MEDLINE

ACCESSION NUMBER:	85177621 MEDLINE
DOCUMENT NUMBER:	85177621 PubMed ID: 3921297
TITLE:	4-Methylumbelliferyl **alpha-N-acetylglucosaminidase** activity for diagnosis of Sanfilippo B disease.
AUTHOR:	Marsh J; Fensom A H
SOURCE:	CLINICAL GENETICS, **(1985 Mar)** 27 (3) 258-62. Journal code: 0253664. ISSN: 0009-9163.
PUB. COUNTRY:	Denmark
DOCUMENT TYPE:	Journal; Article; (JOURNAL ARTICLE)
LANGUAGE:	English
FILE SEGMENT:	Priority Journals
ENTRY MONTH:	198506
ENTRY DATE:	Entered STN: 19900320
	Last Updated on STN: 19900320
	Entered Medline: 19850606

AB Conditions for assay of **alpha-N-acetylglucosaminidase** activity in human cultured fibroblasts, cultured amniotic fluid cells, leucocytes, serum, plasma and chorionic villi were studied using the fluorogenic substrate 4-methylumbelliferyl-2-acetamido-2-deoxy-alpha-D-glucopyranoside. The substrate was found to have advantage both in terms of sensitivity and ease of use over previously-used colorimetric substrates for assay of the enzyme in these tissues, and for diagnosis of Sanfilippo B disease and identification of carriers. It should have particular application in first trimester prenatal diagnosis using chorionic villus biopsies.

=> d l5 ibib ab 1-4

L5 ANSWER 1 OF 4 CAPLUS COPYRIGHT 2002 ACS
ACCESSION NUMBER: 1993:423458 CAPLUS
DOCUMENT NUMBER: 119:23458
TITLE: Characterization of UDP-N-acetylglucosamine:glycoprotein N-acetylglucosamine-1-phosphotransferase from Acanthamoeba castellanii
AUTHOR(S): Ketcham, Catherine M.; Kornfeld, Stuart
CORPORATE SOURCE: Sch. Med., Washington Univ., St. Louis, MO, 63110, USA
SOURCE: Journal of Biological Chemistry (**1992**), 267(16), 11654-9
CODEN: JBCHA3; ISSN: 0021-9258
DOCUMENT TYPE: Journal
LANGUAGE: English

AB The kinetic properties of UDP-N-acetylglucosamine-glycoprotein N-acetylglucosamine-1-phosphotransferase (I) partially **purified** from A. castellanii were studied. I phosphorylated the lysosomal enzymes, uteroferrin (acid phosphatase) and cathepsin D, 3-90-fold better than nonlysosomal glycoproteins and 16-83-fold better than a Man9GlcNAc oligosaccharide. Deglycosylated uteroferrin was a potent competitive inhibitor of the phosphorylation of intact uteroferrin (Ki = 48 .mu.M) but did not inhibit the phosphorylation of RNase B or the simple sugar, .alpha.-methylmannoside. Deglycosylated RNase A did not inhibit the phosphorylation of RNase B or uteroferrin. The results indicated that **purified** I recognizes a protein domain present on lysosomal enzymes but absent in most nonlysosomal glycoproteins. I also exhibited a marked preference for oligosaccharides contg. mannose .alpha.1,2-mannose sequences, but this could not account for the high-affinity binding to lysosomal enzymes. A. castellanii exts. did not contain detectable levels of N-acetylglucosamine-1-phosphodiester .**alpha**.-**N-acetylglucosaminidase**, the 2nd enzyme in the biosynthetic pathway for the mannose 6-phosphate recognition marker. Thus, A. castellanii does not utilize the phosphomannosyl sorting pathway despite expression of very high levels of I.

L5 ANSWER 2 OF 4 SCISEARCH COPYRIGHT 2002 ISI (R)
ACCESSION NUMBER: 91:161070 SCISEARCH
THE GENUINE ARTICLE: FC289
TITLE: SANFILIPPO-B DISEASE - A REEXAMINATION OF A PARTICULAR SIBSHIP AFTER 12 YEARS
AUTHOR: DINATALE P (Reprint)
CORPORATE SOURCE: NAPLES UNIV, FAC MED & CHIRURG, DIPARTIMENTO BIOCHIM & BIOTECNOL MED, VIA SERGIO PANSINI 5, I-80131 NAPLES, ITALY (Reprint)
COUNTRY OF AUTHOR: ITALY
SOURCE: JOURNAL OF INHERITED METABOLIC DISEASE, (**1991**) Vol. 14, No. 1, pp. 23-28.
DOCUMENT TYPE: Article; Journal
FILE SEGMENT: LIFE
LANGUAGE: ENGLISH
REFERENCE COUNT: 21

ABSTRACT IS AVAILABLE IN THE ALL AND IALL FORMATS

AB A particular sibship, with mild and severe types of Sanfilippo B disease within the same family, was re-examined after 12 years. The phenotypes of the mild and of the severe patients were maintained, specifically the mental retardation. Cultures of lymphoblasts from the mild patient were established and proteins were electrophoresed in native conditions and then immunoblotted with specific antibody. Two bands of 182000 and 131000 Da were found, comigrating with the enzyme from normal lymphoblasts and the enzyme from normal urine. The data are discussed in relationship to the molecular defect underlying **alpha-N-acetylglucosaminidase** deficiency and to the ability of the

=> d l6 ibib ab 1-17

L6 ANSWER 1 OF 17 PASCAL COPYRIGHT 2002 INIST-CNRS. ALL RIGHTS RESERVED.
ACCESSION NUMBER: 1994-0110001 PASCAL
COPYRIGHT NOTICE: Copyright .COPYRGT. 1994 INIST-CNRS. All rights reserved.
TITLE (IN ENGLISH): Identification of GM2-gangliosidosis B1 **variant** carriers
AUTHOR: RIBEIRO M. G.; PINTO R.; OLIVEIRA P.; SA MIRANDA M. C.
CORPORATE SOURCE: Univ. Minho, dep. producao sistemas, 4700 Braga, Portugal
SOURCE: Journal of inherited metabolic disease, **(1993)**, 16(6), 1003-1011, 17 refs.
ISSN: 0141-8955 CODEN: JIMDDP
DOCUMENT TYPE: Journal
BIBLIOGRAPHIC LEVEL: Analytic
COUNTRY: Netherlands
LANGUAGE: English
AVAILABILITY: INIST-18251, 354000023933130120
AB GM2-gangliosidosis B1 **variant**, considered a rare disorder with a wide geographical and ethnic distribution, appears to be exceptionally frequent in Portugal. In order to establish a carrier detection method for this disease we have determined the ratio of enzymatic activities against 4MUGS and 4MUG in urine from B1 **variant** obligate carriers and controls, using the total extract and the Hex A immunobound to a monoclonal antibody. The Hex A immunoassay was applied to the identification of carriers in B1 **variant** families and the results obtained were compared with those from DNA analysis. The reliability and feasibility of the Hex A immunoassay make it a suitable method for B1 **variant** carrier screening, which is particularly important for the prevention of this severe neurological disease in the population at risk

L6 ANSWER 2 OF 17 MEDLINE
ACCESSION NUMBER: 94172997 MEDLINE
DOCUMENT NUMBER: 94172997 PubMed ID: 8127069
TITLE: A fluorimetric enzyme assay for the diagnosis of Sanfilippo disease type D (MPS IIID).
AUTHOR: He W; Voznyi YaV; Boer A M; Kleijer W J; van Diggelen O P
CORPORATE SOURCE: Department of Clinical Genetics, Erasmus University, Rotterdam, The Netherlands.
SOURCE: JOURNAL OF INHERITED METABOLIC DISEASE, **(1993)** 16 (6) 935-41.
Journal code: 7910918. ISSN: 0141-8955.
PUB. COUNTRY: Netherlands
DOCUMENT TYPE: Journal; Article; (JOURNAL ARTICLE)
LANGUAGE: English
FILE SEGMENT: Priority Journals
ENTRY MONTH: 199404
ENTRY DATE: Entered STN: 19940420
Last Updated on STN: 19960129
Entered Medline: 19940411
AB 4-Methylumbelliferyl-alpha-N-acetylglucosamine 6-sulphate was synthesized and shown to be a substrate for the lysosomal N-acetylglucosamine-6-sulphate sulphatase (GlcNAc-6S sulphatase). Fibroblasts and leukocytes from 3 different Sanfilippo D patients showed < 1% of mean normal GlcNAc-6S sulphatase activity. The enzymatic liberation of the fluorochrome from 4-methyl-umbelliferyl-alpha-N-acetylglucosamine 6-sulphate requires the sequential action of the GlcNAc-6S sulphatase and **alpha-N-acetylglucosaminidase**. A normal level of **alpha-N-acetylglucosaminidase** activity was insufficient to complete the hydrolysis of the reaction intermediate 4-methylumbelliferyl-alpha-N-acetylglucosaminide formed by the GlcNAc-6S

sulphatase. A second incubation in the presence of excess **alpha-N-acetylglucosaminidase** is needed to avoid underestimation of the GlcNAc-6S sulphatase activity.

L6 ANSWER 3 OF 17 CAPLUS COPYRIGHT 2002 ACS
ACCESSION NUMBER: 1993:423458 CAPLUS
DOCUMENT NUMBER: 119:23458
TITLE: Characterization of UDP-N-acetylglucosamine:glycoprotein N-acetylglucosamine-1-phosphotransferase from Acanthamoeba castellanii
AUTHOR(S): Ketcham, Catherine M.; Kornfeld, Stuart
CORPORATE SOURCE: Sch. Med., Washington Univ., St. Louis, MO, 63110, USA
SOURCE: Journal of Biological Chemistry (**1992**), 267(16), 11654-9
CODEN: JBCHA3; ISSN: 0021-9258
DOCUMENT TYPE: Journal
LANGUAGE: English

AB The kinetic properties of UDP-N-acetylglucosamine-glycoprotein N-acetylglucosamine-1-phosphotransferase (I) partially purified from A. castellanii were studied. I phosphorylated the lysosomal enzymes, uteroferrin (acid phosphatase) and cathepsin D, 3-90-fold better than nonlysosomal glycoproteins and 16-83-fold better than a Man9GlcNAc oligosaccharide. Deglycosylated uteroferrin was a potent competitive inhibitor of the phosphorylation of intact uteroferrin (Ki = 48 .mu.M) but did not inhibit the phosphorylation of RNase B or the simple sugar, .alpha.-methylmannoside. Deglycosylated RNase A did not inhibit the phosphorylation of RNase B or uteroferrin. The results indicated that purified I recognizes a protein domain present on lysosomal enzymes but absent in most nonlysosomal glycoproteins. I also exhibited a marked preference for oligosaccharides contg. mannose .alpha.1,2-mannose sequences, but this could not account for the high-affinity binding to lysosomal enzymes. A. castellanii exts. did not contain detectable levels of N-acetylglucosamine-1-phosphodiester .**alpha**.-**N-acetylglucosaminidase**, the 2nd enzyme in the biosynthetic pathway for the mannose 6-phosphate recognition marker. Thus, A. castellanii does not utilize the phosphomannosyl sorting pathway despite expression of very high levels of I.

L6 ANSWER 4 OF 17 EMBASE COPYRIGHT 2002 ELSEVIER SCI. B.V.DUPLICATE 1
ACCESSION NUMBER: 91107182 EMBASE
DOCUMENT NUMBER: 1991107182
TITLE: Sanfilippo B disease: A re-examination of a particular sibship after 12 years.
AUTHOR: Di Natale P.
CORPORATE SOURCE: Dipartimento di Biochimica e Biotecnologia Mediche, II Facolta di Medicina e Chirurgia, Universita degli Studi di Napoli Federico II, Via Sergio Pansini 5, 80131 Naples, Italy
SOURCE: Journal of Inherited Metabolic Disease, (1991) 14/1 (23-28).
ISSN: 0141-8955 CODEN: JIMDDP
COUNTRY: United Kingdom
DOCUMENT TYPE: Journal; Article
FILE SEGMENT: 007 Pediatrics and Pediatric Surgery
022 Human Genetics
029 Clinical Biochemistry
LANGUAGE: English
SUMMARY LANGUAGE: English

AB A particular sibship, with mild and severe types of Sanfilippo B disease within the same family, was re-examined after 12 years. The phenotypes of the mild and of the severe patients were maintained, specifically the mental retardation. Cultures of lymphoblasts from the mild patient were established and proteins were electrophoresed in native conditions and then immunoblotted with specific antibody. Two bands of 182,000 and

131,000 Da were found, comigrating with the enzyme from normal lymphoblasts and the enzyme from normal urine. The data are discussed in relationship to the molecular defect underlying .**alpha**.-**N-acetylglucosaminidase** deficiency and to the ability of the antiserum to react with normal, **mutant**, monomeric and multimeric forms of the enzyme.

L6 ANSWER 5 OF 17 SCISEARCH COPYRIGHT 2002 ISI (R)
ACCESSION NUMBER: 91:161070 SCISEARCH
THE GENUINE ARTICLE: FC289
TITLE: SANFILIPPO-B DISEASE - A REEXAMINATION OF A PARTICULAR SIBSHIP AFTER 12 YEARS
AUTHOR: DINATALE P (Reprint)
CORPORATE SOURCE: NAPLES UNIV, FAC MED & CHIRURG, DIPARTIMENTO BIOCHIM & BIOTECNOL MED, VIA SERGIO PANSINI 5, I-80131 NAPLES, ITALY (Reprint)
COUNTRY OF AUTHOR: ITALY
SOURCE: JOURNAL OF INHERITED METABOLIC DISEASE, (**1991**) Vol. 14, No. 1, pp. 23-28.
DOCUMENT TYPE: Article; Journal
FILE SEGMENT: LIFE
LANGUAGE: ENGLISH
REFERENCE COUNT: 21

ABSTRACT IS AVAILABLE IN THE ALL AND IALL FORMATS

AB A particular sibship, with mild and severe types of Sanfilippo B disease within the same family, was re-examined after 12 years. The phenotypes of the mild and of the severe patients were maintained, specifically the mental retardation. Cultures of lymphoblasts from the mild patient were established and proteins were electrophoresed in native conditions and then immunoblotted with specific antibody. Two bands of 182000 and 131000 Da were found, comigrating with the enzyme from normal lymphoblasts and the enzyme from normal urine. The data are discussed in relationship to the molecular defect underlying **alpha-N-acetylglucosaminidase** deficiency and to the ability of the antiserum to react with normal, **mutant**, monomeric and multimeric forms of the enzyme.

L6 ANSWER 6 OF 17 MEDLINE
ACCESSION NUMBER: 85177621 MEDLINE
DOCUMENT NUMBER: 85177621 PubMed ID: 3921297
TITLE: 4-Methylumbelliferyl **alpha-N-acetylglucosaminidase** activity for diagnosis of Sanfilippo B disease.
AUTHOR: Marsh J; Fensom A H
SOURCE: CLINICAL GENETICS, **(1985 Mar)** 27 (3) 258-62. Journal code: 0253664. ISSN: 0009-9163.
PUB. COUNTRY: Denmark
DOCUMENT TYPE: Journal; Article; (JOURNAL ARTICLE)
LANGUAGE: English
FILE SEGMENT: Priority Journals
ENTRY MONTH: 198506
ENTRY DATE: Entered STN: 19900320
Last Updated on STN: 19900320
Entered Medline: 19850606

AB Conditions for assay of **alpha-N-acetylglucosaminidase** activity in human cultured fibroblasts, cultured amniotic fluid cells, leucocytes, serum, plasma and chorionic villi were studied using the fluorogenic substrate 4-methylumbelliferyl-2-acetamido-2-deoxy-alpha-D-glucopyranoside. The substrate was found to have advantage both in terms of sensitivity and ease of use over previously-used colorimetric substrates for assay of the enzyme in these tissues, and for diagnosis of Sanfilippo B disease and identification of carriers. It should have particular application in first trimester prenatal diagnosis using chorionic villus biopsies.

L6 ANSWER 7 OF 17 EMBASE COPYRIGHT 2002 ELSEVIER SCI. B.V.DUPLICATE 2
ACCESSION NUMBER: 82174669 EMBASE
DOCUMENT NUMBER: 1982174669
TITLE: Sanfilippo B syndrome (MPS III B): Altered residual . **alpha**.-**N-acetylglucosaminidase** activity in an unusual sibship.
AUTHOR: Di Natale P.; Murino P.; Pontarelli G.; et al.
CORPORATE SOURCE: Ist. Biochim. Cell. Mol., Clin. Pediatr., II Med. Sch., Univ. Naples, 80131 Naples, Italy
SOURCE: Clinica Chimica Acta, (1982) 122/2 (135-143).
CODEN: CCATAR
COUNTRY: Netherlands
DOCUMENT TYPE: Journal
FILE SEGMENT: 029 Clinical Biochemistry
022 Human Genetics
007 Pediatrics and Pediatric Surgery
LANGUAGE: English
AB We studied the residual .**alpha**.-**N-acetylglucosaminidase** activity in two siblings with severe and mild Sanfilippo B syndrome. No striking differences were demonstrated between the **mutant** enzymes from the severe and the mild case. However we found an altered enzyme activity characterized by displacement of the pH optimum toward basic values compared to the pH optimum of the normal enzyme, higher stability to heat and to Hg2+ ion treatment. It is suggested that the Sanfilippo B disease in this sibship is due to a mutation of a structural gene coding for .**alpha**.-**N-acetylglucosaminidase**.

L6 ANSWER 8 OF 17 CAPLUS COPYRIGHT 2002 ACS DUPLICATE 3
ACCESSION NUMBER: 1982:83567 CAPLUS
DOCUMENT NUMBER: 96:83567
TITLE: Leishmania donovani-macrophage binding mediated by surface glycoproteins/antigens: characterization in vitro by a radioisotopic assay
AUTHOR(S): Chang, Kwang Poo
CORPORATE SOURCE: Lab. Parasitol., Rockefeller Univ., New York, NY, 10021, USA
SOURCE: Molecular and Biochemical Parasitology (**1981**), 4(1-2), 67-76
CODEN: MBIPDP; ISSN: 0166-6851
DOCUMENT TYPE: Journal
LANGUAGE: English
AB A radioisotopic assay was developed to quantitate the binding of L. donovani promastigotes to hamster peritoneal macrophages in vitro. The binding was temp. dependent and required no serum factors. Binding was reduced by preloading macrophages with zymosan granules or unlabeled promastigotes, but not with latex beads or opsonized erythrocytes. Binding was reduced by 10 mM EGTA that was reversible by the addn. of an equimolar concn. of Ca2+, but not Mg2+. Sialic acid, D-glucose, D-mannose and their **derivs**. reduced the binding, whereas L-fucose, D-galactose and their related sugars did not. Pretreatment of promastigotes with neuraminidase, .alpha.-mannosidase, .**alpha**.-**N-acetylglucosaminidase** of .beta.-glucosidase reduced their binding to macrophages. Prior trypsinization of either macrophages or promastigotes also reduced the binding. At 4.degree., prior opsonization of promastigotes with subagglutination titers of antiserum doubled the level of binding but in combination with protein A reduced it to 50% of its normal binding level. Prior opsonization of macrophages decreased their binding to promastigotes at 4 or 37.degree.. The results indicate that binding of L. donovani promastigotes to hamster peritoneal macrophages is a ligand-receptor interaction involving their antigenic surface membrane proteins. The binding ligands of the parasites appear to have at least sialic acid, glucosyl, mannosyl and N-acetylglucosaminyl

terminal residues as binding determinants. Thus, receptor-mediated endocytosis, defined in a broader sense, appears to be the mechanism by which leishmanias gain entry into macrophages.

L6 ANSWER 9 OF 17 CAPLUS COPYRIGHT 2002 ACS
ACCESSION NUMBER: 1980:3682 CAPLUS
DOCUMENT NUMBER: 92:3682
TITLE: Inhibition of lysosomal enzyme endocytosis by carbohydrate and lectins
AUTHOR(S): Von Figura, Kurt; Ullrich, Kurt; Mersmann, Guenther; Beeck, Hannelora; Weber, Ernst; Strecker, Gerard
CORPORATE SOURCE: USA
SOURCE: Glycocojugate Res., Proc. Int. Symp., 4th (**1979**), Meeting Date 1977, Volume 2, 951-3. Editor(s): Gregory, John D.; Jeanloz, Roger W. Academic: New York, N. Y.
CODEN: 41RSAU
DOCUMENT TYPE: Conference
LANGUAGE: English
AB Lysosomal enzyme endocytosis by fibroblasts and liver epithelium occurs by binding to cell surface receptors which can also be recognized by specific saccharides, saccharide **derivs**., and lectins. Adsorptive endocytosis of lysosomal .**alpha**.-**N-acetylglucosaminidase**; .beta.-N-acetylglucosaminidase, arylsulfatase A, and .alpha.-mannosidase was specifically and competitively inhibited by D-mannose, L-fucose, Me .alpha.-D-mannopyranoside, p-nitrophenyl .alpha.-glycosides of D-mannose and L-fucose, D-lyxose, D-arabinoside, and mannose 6-phosphate, all of which exerted inhibition by interaction with the cell surface receptor. On treatment of the lysosomal enzymes with alk. phosphatase adsorptive endocytosis was inhibited or moderated for both fibroblasts and liver epithelium cells, indicating that the cell surface receptor recognizes a phosphorylated carbohydrate on lysosomal enzymes. .beta.-Glucuronidase accumulation, the uptake of which was not affected by sugars, was not inhnbited by alk. phosphatase treatment. On pretreatment of fibroblasts with concanavalin A and wheat germ agglutinin, nonspecific inhibition of enzyme endocytosis was obsd. This probably results from the effect of lectins on the lateral mobility of cell surface receptor components. Apparently, the receptor is a glycoprotein and(or) closely coupled to a lectin receptor.

L6 ANSWER 10 OF 17 CAPLUS COPYRIGHT 2002 ACS DUPLICATE 4
ACCESSION NUMBER: 1979:3963 CAPLUS
DOCUMENT NUMBER: 90:3963
TITLE: Sanfilippo syndrome type C: Deficiency of acetyl-CoA:.alpha.-glucosaminide N-acetyltransferase in skin fibroblasts
AUTHOR(S): Klein, Udo; Kresse, Hans; Von Figura, Kurt
CORPORATE SOURCE: Inst. Physiol. Chem., Muenster, Ger.
SOURCE: Proc. Natl. Acad. Sci. U. S. A. (**1978**), 75(10), 5185-9
CODEN: PNASA6; ISSN: 0027-8424
DOCUMENT TYPE: Journal
LANGUAGE: English
AB In fibroblast homogenates from 3 patients with Sanfilippo syndrome type C (mucopolysaccharidosis III C), a biochem. **variant** of the Sanfilippo syndrome, complete deficiency of the acetyl CoA:.alpha.-glucosaminide N-acetyltransferase activity was detected. Activities of all lysosomal hydrolases known so far to degrade mucopolysaccharides, including those of sulfamidase and .**alpha**.-**N-acetylglucosaminidase**, were in the range of controls. Acetyl CoA:.alpha.-glucosaminide N-acetyltransferase activity was normal in fibroblasts of patients with other genetic mucopolysaccharidoses, including Sanfilippo syndrome A and B.

L6 ANSWER 11 OF 17 CAPLUS COPYRIGHT 2002 ACS
ACCESSION NUMBER: 1976:103375 CAPLUS
DOCUMENT NUMBER: 84:103375
TITLE: Sanfilippo disease type B: presence of material cross reacting with antibodies against .**alpha**.-**N-acetylglucosaminidase**
AUTHOR(S): Von Figura, Kurt; Kresse, Hans
CORPORATE SOURCE: Inst. Physiol. Chem., Univ. Muenster, Muenster, Ger.
SOURCE: Eur. J. Biochem. (**1976**), 61(2), 581-8
CODEN: EJBCAI
DOCUMENT TYPE: Journal
LANGUAGE: English

AB .**alpha**.-**N-acetylglucosaminidase** (I) (enzyme deficient in Sanfilippo disease type B) was purified from normal human urine. An antiserum was raised in rabbits against the purified enzyme. Preincubation of the antiserum with crude I from normal human urine, followed by centrifugation, led to a marked redn. of the I activity in the supernatant. Formation of the antibody-enzyme complex had no influence on the activity. The thermal stability of the enzyme was markedly enhanced by complex formation with the antiserum. In the urine from three patients with Sanfilippo disease type B the presence of cross-reacting material could be demonstrated by incubating the antiserum with I in the presence of Sanfilippo B urine or by pretreatment of the antiserum with Sanfilippo B urine. Immunodiffusion and immunoelectrophoresis of crude normal or Sanfilippo B urine gave rise to up to four pptn. lines, only one of which exhibited I activity in the case of normal urine. Purified I yielded only a single pptn. line. After adsorption with the purified enzyme the antiserum did not cross react with any of the urinary proteins. On a quant. detn. of cross-reacting material using Sepharose-immobilized antibodies in the urine from two Sanfilippo B patients the amt. of cross-reacting material appeared to be less than one fourth of the amt. of protein in an age-matched control urine. The cross-reacting material present in the urine of Sanfilippo B patients had a significantly lower binding affinity for antibodies against I than prepns. from normal human urine. It could be calcd. that the amt. of cross-reacting material in the urine of Sanfilippo B patients exceeded that of normal controls. It is concluded that Sanfilippo disease type B is due to a mutation of a structural gene coding for I. The mutation affects the catalytic and immunol. properties of the enzyme protein.

L6 ANSWER 12 OF 17 EMBASE COPYRIGHT 2002 ELSEVIER SCI. B.V.
ACCESSION NUMBER: 77183386 EMBASE
DOCUMENT NUMBER: 1977183386
TITLE: The toxicity of dimethoxyphenol and related compounds in the cat.
AUTHOR: Miller J.J.; Powell G.M.; Olavesen A.H.; Curtis C.G.
CORPORATE SOURCE: Dept. Biochem., Univ. Coll., Cardiff, United Kingdom
SOURCE: Toxicology and Applied Pharmacology, (1976) 38/1 (47-57).
CODEN: TXAPA
DOCUMENT TYPE: Journal
FILE SEGMENT: 037 Drug Literature Index
030 Pharmacology
035 Occupational Health and Industrial Medicine
LANGUAGE: English

AB The metabolic fate of 2,6 dimethoxyphenol, phenol and quinol (hydroquinone) were investigated in the cat. The nature of the urinary metabolic products was dependent upon the dose of the phenol administered, although in all cases the major detoxication products were sulfate conjugates. Hydroxylation of 2,6 dimethoxyphenol and phenol to the corresponding quinols is a major pathway and at relatively high doses unconjugated quinols were found in the urine. Experiments with para substituted phenols suggest that quinol formation is an obligatory step leading to poisoning in the cat. 2,6 Dimethoxyquinol and quinol had no

effect on mitochondrial respiration in vitro whereas the corresponding quinones were potent inhibitors. Inhibition was not observed in the presence of L cysteine.

L6 ANSWER 13 OF 17 EMBASE COPYRIGHT 2002 ELSEVIER SCI. B.V.
ACCESSION NUMBER: 75146724 EMBASE
DOCUMENT NUMBER: 1975146724
TITLE: Inhibition of pinocytosis by cytochalasin B. Decrease in intracellular lysosomal enzyme activities and increased storage of glycosaminoglycans.
AUTHOR: Von Figura K.; Kresse H.
CORPORATE SOURCE: Inst. Physiol. Chem., Univ. Munster, Germany
SOURCE: European Journal of Biochemistry, (1974) 48/2 (357-363). CODEN: EJBCAI
DOCUMENT TYPE: Journal
FILE SEGMENT: 029 Clinical Biochemistry
LANGUAGE: English
AB The influence of cytochalasin B on the pinocytosis of lysosomal enzymes and on the intracellular accumulation, secretion and uptake of sulfated glycosaminoglycans was studied in cultivated skin fibroblasts. The uptake of .alpha. N acetylglycosaminidase was measured in Sanfilippo B fibroblasts, that of .beta. N acetylhexosaminidase in Sandhoff fibroblasts and that of .beta. glucuronidase in fibroblasts from a patient with .beta. glucuronidase deficiency. Cytochalasin B reduces drastically the uptake of these glycosidases. For .**alpha. N acetylglucosaminidase** a dose response relationship and the time interval between application of the drug and the onset of inhibition of pinocytosis are given. When normal fibroblasts are incubated in the presence of cytochalasin b the cells become depleted of the intracellular activity of lysosomal hydrolases but not of the cytoplasmic enzyme lactate dehydrogenase. In the medium and increase of .beta. N acetylhexosaminidase activity is measurable. The decrease of the activity of intralysosomal enzymes mirrors their intracellular half life as determined in **mutant** cell strains. As a consequence of the lowered hydrolase activity excessive amounts of sulfated glycosaminoglycans are accumulated in normal fibroblasts although the pinocytosis of secreted proteoglycans is markedly diminished. The results support the hypothesis that in fibroblasts lysosomal enzymes are primarily secreted and then reach the lysosomes by adsorptive pinocytosis.

L6 ANSWER 14 OF 17 EMBASE COPYRIGHT 2002 ELSEVIER SCI. B.V.
ACCESSION NUMBER: 75126956 EMBASE
DOCUMENT NUMBER: 1975126956
TITLE: [Comparative study of clinical, radiologic, biochemical and genetic features of Sanfilippo's disease of type A and B. Six cases].
ETUDE COMPARATIVE DES ASPECTS CLINIQUES, RADIOLOGIQUES, BIOCHIMIQUES ET GENETIQUES DE LA MALADIE DE SANFILIPPO DE TYPE A ET DE TYPE B. A PROPOS DE 6 OBSERVATIONS.
AUTHOR: Farriaux J.P.; Dhondt J.L.; Blanckaert D.; et al.
CORPORATE SOURCE: Lab. Rech., Clin. Ped., Cent. Hosp. Reg. Lille, Cite Hosp., Lille, France
SOURCE: Helvetica Paediatrica Acta, (1974) 29/4 (349-370). CODEN: HPAAAE
DOCUMENT TYPE: Journal
FILE SEGMENT: 007 Pediatrics and Pediatric Surgery
014 Radiology
005 General Pathology and Pathological Anatomy
LANGUAGE: French
AB Six cases of Sanfilippo disease, 3 of type A (heparin sulfate sulfatase deficiency) and 3 of type B(N acetyl (a)D glucosaminidase deficiency) are described. A comparative study of the clinical, radiological, biological and biochemical features of types showed no significant differences. It is suggested that the diagnosis of Sanfilippo disease **variants** is

dependent upon metabolic proof: enzyme activity levels, mutual correction of the defect in cultured fibroblasts of types A and B, and sulfate incorporation in cultured fibroblasts of types A and B.

L6 ANSWER 15 OF 17 MEDLINE
ACCESSION NUMBER: 75054959 MEDLINE
DOCUMENT NUMBER: 75054959 PubMed ID: 4215452
TITLE: Physical properties and biological activities of two forms of **alpha-N-acetylglucosaminidase** from bovine spleen.
AUTHOR: Mersmann G; von Figura K; Buddecke E
SOURCE: BIOCHIMICA ET BIOPHYSICA ACTA, **(1974 Sep 11)** 364 (1) 88-96.
Journal code: 0217513. ISSN: 0006-3002.
PUB. COUNTRY: Netherlands
DOCUMENT TYPE: Journal; Article; (JOURNAL ARTICLE)
LANGUAGE: English
FILE SEGMENT: Priority Journals
ENTRY MONTH: 197503
ENTRY DATE: Entered STN: 19900310
Last Updated on STN: 19970203
Entered Medline: 19750319

L6 ANSWER 16 OF 17 EMBASE COPYRIGHT 2002 ELSEVIER SCI. B.V.
ACCESSION NUMBER: 74117718 EMBASE
DOCUMENT NUMBER: 1974117718
TITLE: The metabolism and toxicity of phenols in cats.
AUTHOR: Miller J.J.; Powell G.M.; Olavesen A.H.; Curtis C.G.
CORPORATE SOURCE: Dept. Biochem., Univ. Coll., Cardiff, United Kingdom
SOURCE: Biochemical Society Transactions, (1973) 1/5 (1163-1165).
CODEN: BCSTB5
DOCUMENT TYPE: Journal
FILE SEGMENT: 037 Drug Literature Index
029 Clinical Biochemistry
030 Pharmacology
LANGUAGE: English

L6 ANSWER 17 OF 17 CAPLUS COPYRIGHT 2002 ACS
ACCESSION NUMBER: 1957:81884 CAPLUS
DOCUMENT NUMBER: 51:81884
ORIGINAL REFERENCE NO.: 51:14847h-i,14848a-c
TITLE: Glycosidases in mammalian sperm and seminal plasma
AUTHOR(S): Conchie, J.; Mann, T.
CORPORATE SOURCE: Rowett Research Inst., Aberdeen, UK
SOURCE: Nature **(1957)**, 179, 1190-1
DOCUMENT TYPE: Journal
LANGUAGE: Unavailable
AB The object of the investigation was to extend the study of .beta.-glucuronidase, .beta.-N-acetylglucosaminidase, and .alpha.-mannosidase to sperm, seminal plasma, and male accessory secretions of animals other than the rat and to compare the activities of these glycosidases with those of .beta.-mannosidase, .alpha.-and .beta.-glucosidases, .alpha.-and .beta.-galactosidases, .beta.-xylosidase, and .**alpha**.-**N-acetylglucosaminidase**. The substrates used were phenolphthalein glucuronide and phenol and o- or p-nitrophenol **derivs**. of other glycosides. Results were expressed in units defined as .gamma. of aglycone (phenolphthalein, phenol, and o- and p-nitrophenol, resp.) liberated by 1 ml. semen or accessory-gland secretion in 1 hr. at 37.degree.. The species investigated were the ram, bull, boar, stallion, rabbit, dog, and man. Expts. on ram semen showed that .alpha.-mannosidase and .beta.-N-acetylglucosaminidase, in contrast to the other glycosidases studied, were present in the spermatozoa themselves. Out of 370 units of .alpha.-mannosidase and 20,000 units of .beta.-N-acetylglucosaminidase

found in 1 ml. of ram semen, 320 and 4000 units, resp., were derived from spermatozoa. Procedures which caused structural damage or partial disintegration of sperm cells not only failed to release more .alpha.-mannosidase but produced a definite decrease in activity. Of the remaining glycosidases present in ram semen, .beta.-mannosidase, .beta.-galactosidase, and .beta.-glucuronidase were confined chiefly to the seminal plasma; glucosidases and .alpha.-galactosidase were poorly active and .beta.-xylosidase and **.alpha.-N-acetylglucosaminidase** were absent. Results obtained with ejaculated semen, seminal plasma, and accessory-gland secretions of species other than sheep were presented in a table. The outstanding feature was the extraordinarily high level of .beta.-N-acetylglucosaminidase and .alpha.-mannosidase activities in the epididymal seminal plasma. .beta.-Xylosidase and **.alpha.-N-acetylglucosaminidase** showed negligible activity in all species.